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ABSTRACT

This document presents the consensus statement of a 2-day conference which addressed issues concerned with the early identification of hearing impairment. The conference resulted in the following consensus conclusions: (1) all infants admitted to the neonatal intensive care unit should be screened for hearing loss prior to discharge; (2) universal screening should be implemented for all infants within the first 3 months of life; (3) the preferred model for screening should begin with an evoked otoacoustic emissions test and should be followed by an auditory brainstem response test for all infants who fail the evoked otoacoustic emissions test; (4) comprehensive intervention and management programs must be an integral part of a universal screening program; (5) universal neonatal screening should not be a replacement for ongoing surveillance throughout infancy and early childhood; and (6) education of primary caregivers and primary health care providers on the early signs of hearing impairment is essential. Also included in this document are the recommendations of the National Institute on Deafness and Other Communication Disorders concerning acceptable protocols in statewide universal newborn hearing screening programs. These recommendations urge use of screening which involves a physiologic response implemented with objective response criteria. (DB)

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Recommendations of the NIDCD Working Group on Early Identification of Hearing Impairment on Acceptable Protocols for Use in State-Wide Universal Newborn Hearing Screening Programs

This document was developed as part of the National Institute on Deafness and Other Communication Disorders (NIDCD) Workshop on Universal Newborn Hearing Screening held in Chevy Chase, Maryland on September 4-5, 1997. Members of the Working Group on Early Identification of Hearing Impairment in attendance at this workshop and who participated in the writing of this document included: G. Pamela Burch-Sims, Ph.D., Tennessee State University; Richard A. Chole, M.D., Ph.D., University of California; Allan Diefendorf, Ph.D., Indiana University School of Medicine; Karen Doyle, M.D., Ph.D., University of California at Irvine; Stephen Epstein, M.D., Wheaton, Maryland; Judith Gravel, Ph.D., Albert Einstein College of Medicine; Deborah Hayes, Ph.D., Children's Hospital, Denver; Linda Hood, Ph.D., Louisiana State University Medical Center; Susan Jerger, Ph.D., The University of Texas at Dallas; Mary Pat Moeller, M.S., Boys Town National Research Hospital; Susan Norton, Ph.D., Children's Hospital and Medical Center; Beth Prieve, Ph.D., Syracuse University; Patricia Stelmachowicz, Ph.D., Boys Town National Research Hospital; Judith Widen, Ph.D., University of Kansas Medical Center; and Christine Yoshinaga-Itano, Ph.D., University of Colorado.

I. Introduction

The early identification of children with hearing impairment is an important public health objective in the United States. Each year in this country, approximately 1.5 to 3 per 1,000 children are born with significant hearing impairment. With an annual birth rate of approximately 4 million infants, this prevalence rate translates into as many as 33 children per day born with hearing impairment. Currently, many of these children are not identified until the second year of life or later despite advances in the technology available for the early detection of hearing impairment. This delay in identification contrasts with some available statistics from other developed countries where the age of identification has been reduced to less than one year of age. The consequences of a late diagnosis of hearing impairment are significant delays in spoken language and literacy. Without appropriate and timely identification and intervention, early childhood hearing impairment interferes with the development of oral/aural communication, impedes academic performance, and results in long-term vocational consequences.

A National Institutes of Health Consensus Conference held in March of 1993 recommended hearing screening of all newborns, termed universal newborn hearing screening. Access to the largest possible number of newborns is necessary to promote early identification of hearing impairment for all infants and subsequent referral for diagnosis and intervention. The best opportunity for achieving this goal appears to be provided by the development of hearing screening programs for newborns in hospital nurseries or in birthing centers, prior to discharge. The successful implementation of this

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pro-active approach should lead to a greater likelihood that a child with hearing impairment will enjoy academic, social, and vocational success. Recent data indicate that the direct cost of universal newborn hearing screening programs is comparable to the direct cost of universal screening programs for other congenital conditions such as hypothyroidism, phenylketonuria (PKU), and hemoglobinopathies.

A variety of current technologies are available to identify hearing impairment in the first days of life. Two of the current methodologies generally established as effective for universal newborn hearing screening are auditory brainstem responses (ABR) and evoked otoacoustic emissions (EOAE). The focus of this document is to recommend acceptable procedures that can be used by States for universal newborn hearing screening. It is important to note, however, that a newborn hearing screening program is only one component of a comprehensive approach to the management of childhood hearing impairment. The process also requires follow-up diagnostic services, counseling, intervention programs, and parental educational programs. This comprehensive process must be administered by a multidisciplinary team consisting of individuals such as audiologists, physicians, educators, speech/language pathologists, nurses, and parents.

II. Current Status of Statewide Systems for Universal Newborn Hearing Screening

The development of statewide systems for universal newborn hearing screening has been addressed by three approaches, namely legislative mandate, voluntary initiative, and federal support. Legislative mandates have been used to initiate some form of newborn hearing screening in approximately 20 states. In four of the states (Rhode Island, Hawaii, Colorado, and Mississippi), the legislation specifically addresses the screening of all newborns. In the 16 remaining states, however, the legislation addresses the identification of risk status in infants, which indirectly identifies a pool of infants who are at-risk for hearing impairment and who should be screened. A limitation of this latter approach is that screening programs restricted to infants with risk factors for hearing impairment identify only 50% of infants with significant hearing impairment. Thus, health care professionals in several states are attempting to initiate hearing screening services for all newborns, including neonates without risk factors. With regard to voluntary initiatives, voluntary programs exist in several of the states without legislative mandates. For example, Wyoming has achieved effective universal newborn hearing screening without legislation. Numerous local voluntary programs within individual communities or hospitals also exist. Finally, with regard to federal support, funds from the Bureau of Maternal and Child Health have allowed 17 states to commit to achieving universal hearing screening by the year 2000. When fully operational, these 17 states will screen more than 1,000,000 newborns a year.

In concert with recommendations of NIH/NIDCD and the Joint Committee on Infant Hearing (JCIH)*, current statewide programs generally employ ABR and/or EOAE for hearing screening. These techniques are physiological measures of the status of the peripheral auditory system that are highly correlated with hearing status. The techniques permit the identification of infants with communicatively significant hearing impairment

without referring large numbers of normally hearing infants for unnecessary follow-up testing.

III. Acceptable Protocols

Acceptable hearing screening protocols should have specific response attributes and measurement characteristics. Some of these desired response and methodological features are the following:

- The response should be robust (i.e., capable of being measured reliably under a wide variety of conditions);
- The response should be dependent upon the integrity of the peripheral auditory system;
- The response should have predictive value (i.e., it should be present in nearly all normal hearing infants and abnormal in nearly all infants with hearing impairment);
- The response should be measured non-invasively;
- The procedure should employ scientifically-based, objective criteria to define both the method for a technically correct screening test and the guideline for a "pass versus refer" rule;
- The procedure should be capable of testing each ear independently;
- The procedure should achieve a low referral rate for additional testing at another session to prevent unnecessary costs and parental anxiety;
- The procedure should be manageable in a hospital or birthing center setting in order to provide access to the greatest number of neonates, thus promoting the universality of hearing screening;
- After the initial screening, and before the infant returns for any recommended follow-up screening, acceptable protocols should attain a referral rate of no more than approximately 5% for neonates with no risk indicators and 8% for infants at risk for hearing impairment. Previous research indicates that these referral rates can be achieved in approximately 6 months with appropriate training and quality control monitoring; and
- The choice of a particular method will vary as a function of the demographic characteristics of the neonates to be screened and available community resources.

Based on a review of published data, a physiological response implemented with objective response criteria best meets the above requirements. Acceptable approaches include: 1) auditory brainstem response (ABR); 2) either transient evoked otoacoustic emissions (TEOAE) or distortion product evoked otoacoustic emissions (DPOAE); or, 3) a combination of OAE and ABR. Future research may yield additional objective, physiological measures that could advance universal newborn hearing screening. A variety of non-auditory factors may influence the outcome of any of the current hearing screening approaches. These include test environment, infant state, infant medical status, and age. The skill and commitment of the examiner are also important factors influencing the screening test results. Studies have documented that the actual screening can be carried out effectively by a wide variety of personnel with appropriate training,

such as nurses or volunteers. Training and quality assurance measures are vital components for the efficiency and overall effectiveness of screening programs.

The initial hearing screening should be carried out before 3 months of age to ensure that intervention can begin between birth and 6 months of age. The initial screening should represent only one component of an overall identification and intervention program for children with hearing impairment. The overall screening program should include provisions for: 1) tracking of infants who are referred from the initial screening; 2) follow-up diagnostic testing; and, 3) intervention for those infants with confirmed hearing impairment. These programmatic goals can best be achieved through a multi-disciplinary approach including, but not limited to, primary health care providers, neonatologists, audiologists, otolaryngologists, educators, speech-language pathologists, psychologists, and parents. In addition, an important determiner of the success of universal hearing screening programs is the effective, timely and sensitive communication of the results and of any necessary follow-up recommendations to parents or other legal guardians and to primary care providers.

Finally, because some hearing impairments develop during early childhood, the Working Group recommends that children continue to be monitored for hearing impairment and that all children undergo hearing screening upon entering school and periodically thereafter.

IV. Summary

As many as 12,000 infants are born each year in the United States with hearing impairment. Many of these children are not identified as being hearing impaired until they are 2 years of age or older. A delay in the diagnosis of hearing impairment leads to delays in language acquisition and academic achievement. Early identification of and appropriate intervention for children with hearing impairment leads to improvements in speech and language development in affected children, thereby improving the likelihood of positive social, emotional, cognitive, and academic development. Identification of and intervention for infants prior to 6 months of age seems to result in the most favorable outcomes. Therefore, the Working Group recommends that a system of universal hearing screening within newborn nurseries be instituted. Universal screening can be achieved with low cost methods that successfully differentiate newborns with hearing impairment from newborns with normal hearing. The chosen screening method should be simple and the response should be reproducible; the method should be capable of being administered by a variety of adequately trained personnel. At the present time, the Working Group concurs that one or more of the following screening strategies are suitable for application to infant screening: 1) auditory brainstem response (ABR); 2) either transient evoked otoacoustic emissions (TEOAE) or distortion product evoked otoacoustic emissions (DPOAE); or, 3) a combination of OAE and ABR. Universal screening of newborns will lead to the referral of more infants for diagnosis and intervention. Comprehensive intervention and management programs are an essential part of a universal hearing screening program.

*Footnote 1. JCIH is a multidisciplinary committee composed of representatives from the American Academy of Audiology, American Academy of Otolaryngology-HNS, American Academy of Pediatrics, American Speech-Language-Hearing Association, Council for Education of the Deaf, and Directors of Speech and Hearing Programs in State Health and Welfare Departments.